

Mandated Congenital Cytomegalovirus (cCMV) Testing: The Utah EHDI Experience

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- What is CMV?
- Congenital-CMV and Hearing Loss
- Utah LAW
- Utah EHDI Changes
- Ensuring Buy-In
- Q&A



What is CMV?

- A common virus that is a member of the herpes family
- Affects people of all ages
- Most people who are infected with CMV have no signs or symptoms
- Those that do may experience mild flu-like symptoms

When CMV infection occurs during a woman's pregnancy, the baby can become infected before birth =
Congenital CMV infection





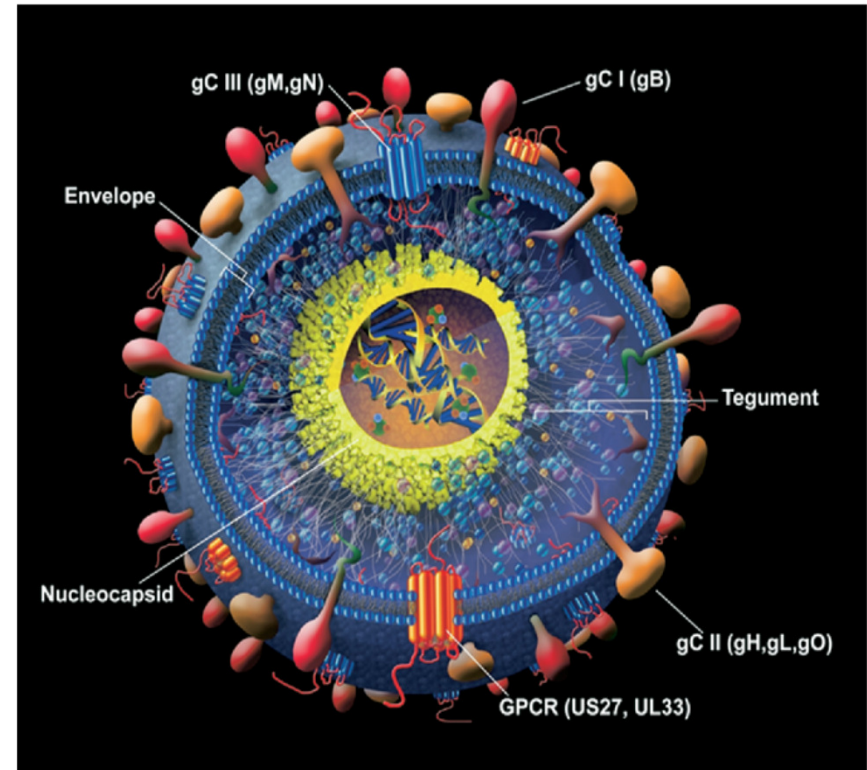
30-40%

About 1 of every 5 children born with congenital CMV will develop permanent problems.....such as **hearing loss** or developmental disabilities, due to the infection.

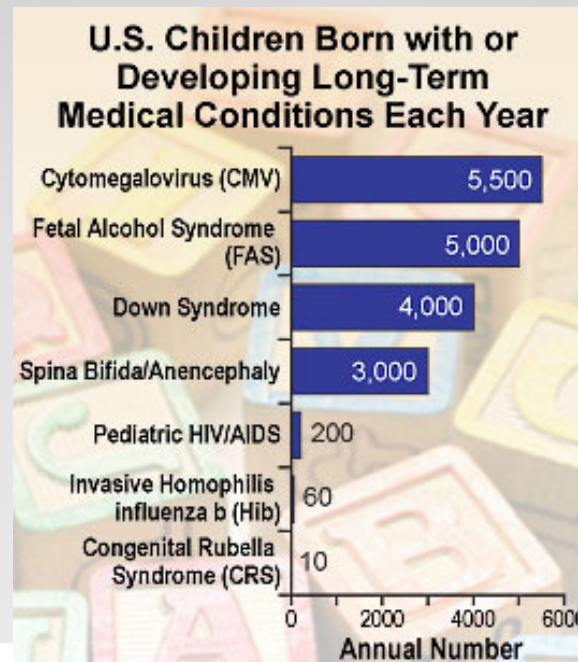


Cytomegalovirus

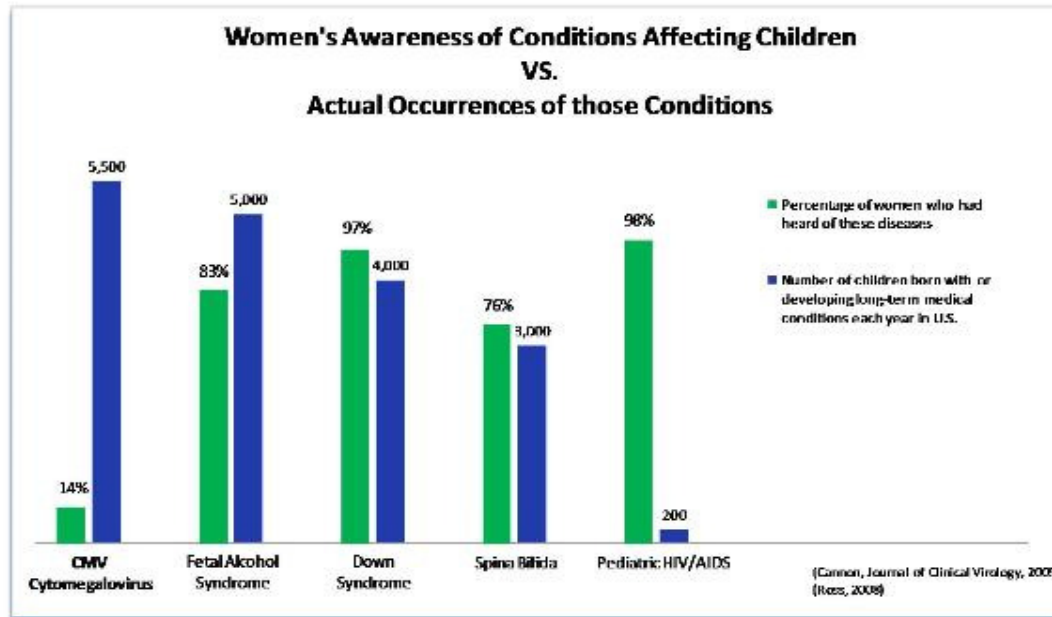
- Cost C-CMV greater than \$ 4 billion/yr in US



Congenital CMV causes more long-term health problems and childhood deaths than Down Syndrome, Fetal Alcohol Syndrome, and Neural Tube Defects (spina bifida, anencephaly).



Compare to CMV Awareness



Brendan B. McGinnis Congenital CMV Foundation
www.cmvfoundation.org
© 2010

“SYMPTOMATIC”:

- 10% fetal demise
- Prematurity

Common features:

Hepatomegaly

Splenomegaly

Petechiae

Jaundice

Microcephaly

Chorioretinitis

Sensorineural hearing loss



“ASYMPTOMATIC”



5-15% of asymptotically infected children will end up having permanent hearing loss.



Congenital CMV is the most common cause of non-hereditary hearing loss in children.

In a large number of asymptomatic children with congenital CMV, hearing loss is the only sequelae (known abnormality).



26-10-10 UCA, “Cytomegalovirus (CMV) **Public Education and Testing**”

- **UDOH establish and conduct a public education program** to inform *pregnant women and women who may become pregnant* about CMV (incidence, transmission, birth defects, diagnostic methods, preventative measures)
- Provide information to: *child care providers, school nurses, health educators, health care providers, religious organizations offering children’s programs as part of worship services*

The virus is generally passed from infected people to others through direct contact with body fluids, such as **urine** or **saliva**.

For pregnant women, one of the most common ways they are exposed to CMV is by **contact with saliva or urine of children who recently had the virus.**



No vac

No FD



26-10-10 UCA, “Cytomegalovirus (CMV) Public Education and **Testing**”

If a newborn infant fails the newborn hearing screening test(s).....

Medical Practitioner shall:

- ***Test the newborn infant for CMV before 21 days of age...unless the parent objects; and***
- ***Provide to the parents information re: birth defects caused by congenital CMV and available methods of treatment.***



Utah EHDI??

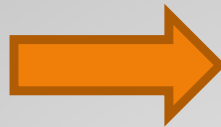
- CDC 1-3-6:

Hearing screening before **1** month

Diagnostic evaluation before **3** months

Early intervention before **6** months





Utah EHDI Goals



- 1/2 - 3 - 6
- 14 DAYS
- 21 DAYS



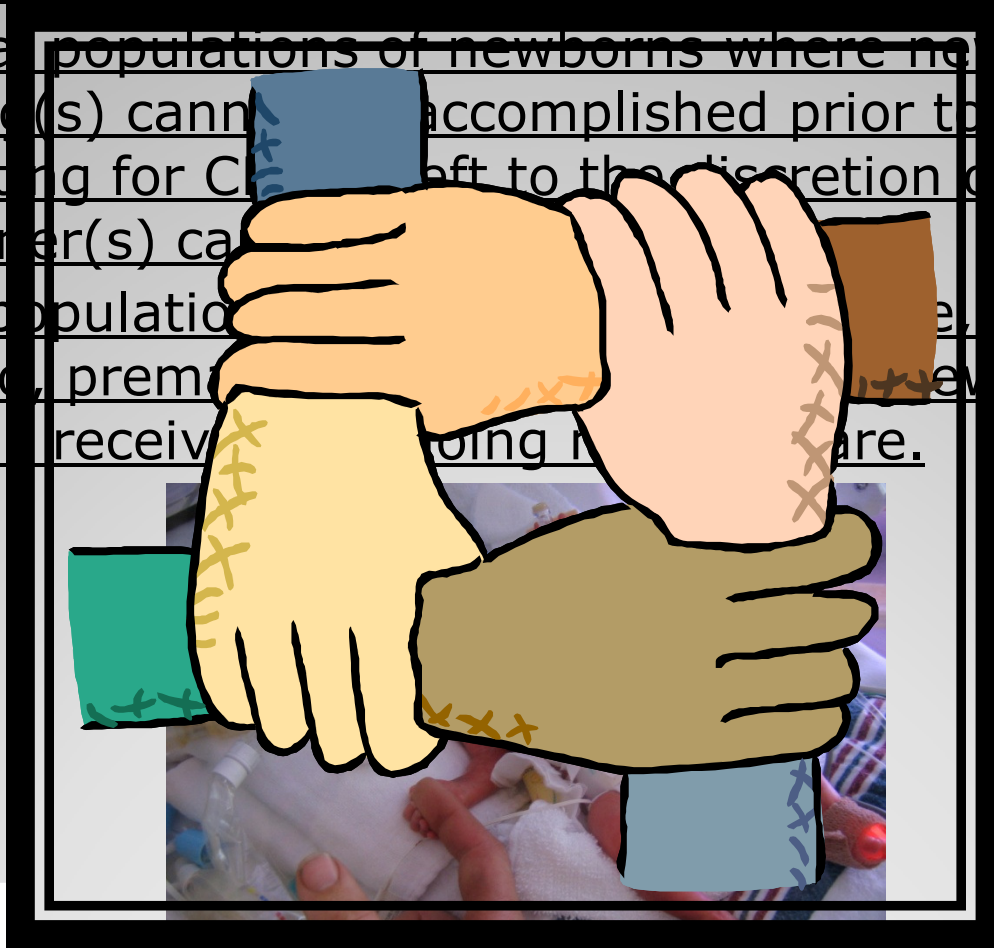
Utah EHDI Goals

- **R398-4-3. Clarification of when a newborn fails a hearing screen.**
- **The newborn must fail both hearing screens, the initial hearing screen routinely done at birth **and** the subsequent follow-up screen **or** if/when the initial failed hearing screen is obtained after 14 days of age before the medical practitioner is required to test for CMV.**



- **R398-4-4. Special populations of newborns.**

- In special populations of newborns where newborn hearing screening(s) cannot be accomplished prior to 21 days of age, testing for C... left to the discretion of the medical practitioner(s) can...
- Special populations include, but not limited to, premature newborns or newborns receiving...



H.B. 81 (2013 General Session) Cytomegalovirus Public Health Initiative (UCA 26-10-10) Sequence of Events

INFANT FAILS INPATIENT NEWBORN HEARING SCREENING (NBHS)

Hospital NBHS Program notifies family their infant failed hearing screen and **schedules outpatient re-screen to take place prior to 14 days of age** emphasizing importance of completing this appointment at the scheduled time.

Hospital NBHS Program obtains complete primary care provider (PCP) information from family and enters it in infant's Hi*Track record. If complete information is not in the physician database "drop down" menu, it can be added/updated by contacting Utah's Early Hearing Detection & Intervention (EHDI) Hi*Track Data Coordinator at (801) 584-8221.

Hospital NBHS Program advises PCP of **initial screen failure** using the "Newborn Hearing Screening / CMV Status Report FAX" form or other mutually agreed upon method of notification. The method of notification must be documented in Hospital Program Summary.

INFANT FAILS 2ND (OUTPATIENT) HEARING SCREENING

Hospital NBHS Program immediately notifies PCP of the **follow-up hearing screening failure**: 1) via fax using the "Newborn Hearing Screening / CMV Status Report FAX" form, 2) phone message, (action must be documented in HiTrack), or 3) other **preferred method** of communication (action must be documented in HiTrack). The method of notification must be documented in Hospital Program Summary. Family is told to contact PCP if does not hear from MD within 24 hours.

PCP discusses congenital CMV with family and orders CMV PCR assay testing via saliva or urine on infant unless parents object.

TESTING OCCURS PRIOR TO 21 DAYS OF AGE

After test results are received, PCP faxes "Newborn Hearing Screening / CMV Status Report FAX" Form to Hospital NBHS Program and to UDOH at (801) 584-8492. PCP consults with family on next steps.

Many babies with hearing loss will react to loud sounds. The only way to know if your baby's hearing is normal is to have the screening done with special equipment.



Before you leave the hospital, be sure you know the results of your baby's hearing screening. If your baby did not pass, he/she should be rescreened before 14 days of age.

What can my baby hear during pregnancy?

- By 7 weeks gestation your baby's ear structures are beginning to develop.
- By 25 weeks gestation your baby will begin to hear and respond to sounds.
- Your speech is one of the predominant uterine sounds that your baby hears. He/she is already learning language in the womb!
- Newborns prefer their mother's voice to that of another female.

CMV AND HEARING LOSS

Cytomegalovirus (CMV) is a common virus that infects people of all ages; however, when it occurs during pregnancy, the baby can become infected potentially causing damage to the brain, eyes and/or inner ears.

For more information go to:

www.health.utah.gov/cshen/chss/cm.html

Facts about newborn hearing screening

Hearing loss is invisible

Most babies with hearing loss have no signs or symptoms. Infants with hearing loss cry and make other sounds just like babies with normal hearing. The first three years are the most important for learning language and how to communicate.

Hearing loss is a very common birth defect

About 3 in every 1000 babies are born with a permanent hearing loss. Your baby needs normal hearing to develop normal speech and language.

How is a hearing screening done?

Your baby will be screened for hearing loss in the hospital soon after birth. The screening takes 10 - 15 minutes while the baby is quiet or asleep. The screening is done with special equipment that is safe and does not hurt.

If your baby does not pass their screening, the hospital will need to notify your pediatrician.

It is important to have your pediatrician chosen BEFORE you have your baby.

Is my baby at risk for hearing loss?

If your baby has **one or more** of the following risk factors or conditions at birth, your baby may be at increased risk for hearing loss:

- Stayed in the NICU for more than 5 days
- Had an infection before or after birth such as Cytomegalovirus (CMV), herpes, rubella or meningitis
- Has a family member with hearing loss from birth or childhood

Babies at risk can pass a hearing screening at birth but will need more testing later. Talk with your baby's doctor and make an appointment with a pediatric audiologist for further testing.

90—95% of children born with permanent hearing loss have normal hearing parents



**Newborn Hearing Screening (NBHS) / CMV
Status Report FAX (1 form with 3 options)**

Date: _____

PILOT

Version:
August 26,
2013
COMPLETE
ONE OPTION
ONLY

TO: _____

FROM: _____ CLINIC/HOSPITAL: _____ FAX: _____ PHONE: _____

OPTION 1 (NBHS Program)

PER H.B. 81 (2013 General Session) UCA 26-10-10 Cytomegalovirus (CMV) Public Health Initiative, the following infant who lists you as their Primary Care Physician, has **FAILED** the **INITIAL** newborn hearing screen and will require a follow-up re-screen. This follow-up appointment is scheduled as noted. Should you have contact with the family prior to the follow-up date please encourage them to keep the appointment as this should be completed **no later than 14 days of age**. Otherwise, no action on your part is necessary at this time.

Notification of failed INITIAL HEARING SCREEN – to be completed by HOSPITAL

| Infant's Name | D.O.B. | Mother's Name | Contact# | Follow-up Appt. |
|---------------|--------|---------------|----------|-----------------|
| | | | | |

OPTION 2 (NBHS Program)

This is to advise the PCP that the following infant has **FAILED** the **FOLLOW-UP (2nd) HEARING SCREENING** and is a candidate for **congenital CMV testing before 21 days of age** per H.B. 81. He/she has also been referred to a pediatric audiologist for a diagnostic hearing evaluation.

Notification of failed FOLLOW-UP HEARING SCREEN – to be completed by HOSPITAL

| Infant's Name | D.O.B. | Mother's Name | Contact# | Diagnostic Appt. |
|---------------|--------|---------------|----------|------------------|
| | | | | |

OPTION 3 (Primary Care Provider)

After congenital CMV PCR assay testing has been completed, PCP office should fax this form back to the Hospital NBHS program at the above listed fax # **AND** to the Utah Department of Health Early Hearing Detection and Intervention (EHDI) program at (801) 584-8492.

Notification of CMV TEST RESULTS – to be completed by PHYSICIAN

| Infant's Name | D.O.B. | Urine (U) or Saliva (S) | RESULT: Pos (+) or Neg (-) |
|---------------|--------|-------------------------|----------------------------|
| | | | |

If questions, please contact the Utah Department of Health Early Hearing Detection and Intervention program at (801) 584-8215.



CMV CONGENITO Y LA PERDIDA DE AUDICION

Citomegalovirus (ci-to-mé-ga-lo-vi-rus), o CMV, es un virus común que infecta a las personas de todas las edades.

La mayoría de las infecciones por CMV son "silenciosas", es decir, la mayoría de las personas que están infectadas con el CMV no presentan signos ni síntomas, y hay ningún efecto perjudicial.

Sin embargo, cuando ocurre el CMV durante el embarazo de una mujer, el bebé puede ser infectado antes de nacer.

La infección por CMV antes del nacimiento se conoce como "**CMV Congénito**". Cuando esto sucede, el virus puede transmitirse al bebé antes de nacer y potencialmente dañar el cerebro, el ojo y/o el oído interno.

Si el oído interno se daña, el bebé puede nacer con pérdida de audición, o desarrollar pérdida de audición después del nacimiento o durante la infancia temprana.



La Detección de Audición del Recién Nacido

Si su bebé no pasa la prueba de audición del recién nacido en el hospital, es muy importante que él tenga otra prueba (pacientes ambulatorios) antes de los 14 días de edad. Para repetir la prueba de audición es necesario completar el proceso de examen de audición del recién nacido, y debe ser programado antes de dado de alta a su bebé.

Otra Prueba de Audición en la Clínica

Si su bebé no pasa la segunda prueba de audición su pediatra o médico debe hablar con usted acerca de las pruebas para su bebé por CMV congénito.

Las Pruebas de CMV congénito son simples y sin dolor y se pueden lograr utilizando una muestra de orina o muestra de saliva (el interior de la mejilla del bebé se limpia para una muestra).

Con el fin de detectar con precisión CMV congénito, **esta prueba de laboratorio debe realizarse en muestras tomadas antes de que su bebé tenga 21 días de edad.**

¿Qué pasa si mi bebé tiene CMV congénito?

El médico guiará la atención médica de su bebé y lo enviará a especialistas que se necesitan.

HECHOS CMV

- La mayoría de los adultos y los niños que están infectados con CMV no sienten enfermos y no saben que han sido infectados; otras pueden tener síntomas leves similares a la gripe
- Los bebés y los niños que están infectados con CMV después del nacimiento casi nunca tienen problemas
- La pérdida de audición por CMV congénita puede afectar a uno o ambos oídos; puede afectar a todos o algunos de los tonos que el bebé está oyendo; puede ser leve o grave; puede presentarse en el nacimiento o aparecer más tarde; y puede ser progresiva (empeorando con el tiempo)
- 50% de los bebés con CMV congénito pasará su prueba de audición del recién nacido, y luego comenzará una pérdida de audición

CMV congénita es la causa más común de pérdida de la audición no hereditaria en los niños



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PILOT

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Notification of failed FOLLOW-UP HEARING SCREEN – to be completed by HOSPITAL

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|---------------|--------|---------------|----------|------------------|
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|---------------|--------|-------------------------|----------------------------|
| | | | |

If questions, please contact the Utah Department of Health Early Hearing Detection and Intervention program at (801) 584-8215.



Additional documents created for this initiative

New EHDI sequence of events

H.B. 81 (2013 GENERAL SESSION)

Congenital CMV and Hearing Loss
brochure for parents

UAC 26-10-10

Utah Early Hearing Detection and Intervention
PROCESS UPDATE

All about CMV

Newborn Hearing Screening / CMV
Status Report fax



H.B. 81 (2013) =
CYTOMEGALOVIRUS
PUBLIC HEALTH
INITIATIVE

Chief Sponsor: Representative Ronda
Menlove

Governor signed into law = UAC 26-10-10

- 1) UDOH Education
- 2) PCPs test infants who fail two
hearing screenings for congenital
CMV



Training the Front Line



WE WANT TO HEAR FROM YOU!

Feedback on.....

How is the new NBHS protocol going for you?

What have primary care providers been saying?

What have families been saying?

What have your screeners been saying?

Fax form?

How can we help?

Utah EHDI Survey December 2013

Please take a few moments to answer the following questions from your own program's perspective (your families/babies):

- 1) What do you feel contributes the most to loss to follow-up between a failed first newborn hearing screen (typically inpatient) and completing a second screen (typically outpatient)?

- 2) What do you feel contributes the most to loss to follow-up between a failed second screen and completion of a diagnostic evaluation?

- 3) What do you feel contributes the most to an eligible child not enrolling in early intervention?

- 4) What problems / comments / feedback have you encountered from stakeholders re: the mandated CMV testing?**

- 5) What contributes the most to loss to documentation?

- 6) If a magic wand could be waved, what has the most potential to decrease your program's loss to f-u rate?



Continued Communication



Form Strong Partnerships

UPDATE: Cytomegalovirus Public Health Initiative & Testing

Stephanie McVicar, Au.D., CCC-A, Director
Utah Early Hearing Detection and Intervention, Utah Department of Health
Email: smcvicar@utah.gov

Data Update

It has been almost six months since UCA 26-10-10, the Cytomegalovirus Public Health Initiative and Testing law, was implemented in Utah. During this time, 25 infants have been referred for CMV testing with results reported to the Utah Department of Health (UDOH). Four of these were positive for congenital CMV. However, according to what has been reported in HI*TRACK, the UDOH Early Hearing Detection and Intervention (EHDI) database, there have been 87 infants eligible for CMV testing, having failed two newborn hearing screenings. For these 62 infants with no CMV test results reported to the UDOH, were the CMV tests not done or just not reported?

Rules Update

The rules for UCA 26-10-10 include reporting requirements for medical practitioners. **Medical practitioners are required to submit results of the CMV testing to UDOH for each newborn under their care that is referred for CMV testing within 10 days of receiving results.** The rules also clarify when a newborn must be referred for CMV testing: if the infant fails both the initial hearing screen routinely done following birth *and* the subsequent follow-up screen *or* when the initial screen is obtained after 14 days of age and the newborn fails it. The rules also state that special populations of newborns do exist where newborn hearing screening(s) cannot be accomplished prior to 21 days of age, and in those situations, testing for CMV is left to the discretion of the medical practitioner(s) caring for the newborn. Special populations of newborns may include, but are not limited to, premature or medically fragile newborns or newborns receiving on-going medical care.

Testing Update

If the infant is 4 weeks of age or younger at the time of CMV testing, PCR testing of saliva is the preferred method to detect CMV, given the ease of saliva sampling in young infants. This should have >95% sensitivity, based on the study of Boppana et al. (New Engl J Med 2011;364:2111-8). *PCR testing of urine for CMV is an acceptable alternative.* Because of their low sensitivity, serologic studies or PCR testing of *blood* for CMV should not be performed.

Another caution of which to be aware is that newborn screening of congenital cytomegalovirus infection using saliva can be influenced by breast feeding. Koyano et al at Asahikawa Medical University in Japan recommend that saliva testing be done **right before a feeding** as their preliminary data has shown that CMV in breast milk can be detected in an infant's saliva for at least 30 minutes after feeding, possibly resulting in a false positive for congenital CMV in the infant.

Worth mentioning again is that the preferred specimen for the ARUP CMV saliva test is saliva collected in the ORAcollect OC-100 kit, and at present, ARUP is the only local facility with a validated test for saliva. Many labs, however, can perform PCR testing on urine. Complete information on CMV testing for UCA 26-10-10 can be found in the **CMV PCR Testing** document on our CMV website at: <http://www.health.utah.gov/cshcn/CHSS/CMV.html>. That and many other items, including a new **CMV Testing Refusal Form**, can be found there.

UDOH Information

The fax # for CMV information to be sent is (801) 584-8492. Primary care providers should receive notification of an infant's failed screening(s) from the newborn hearing screener on the **CMV and NBHS Testing Status FAX Form** which is available on our website. This form is used both for notification of the PC by the hearing screener and for notification of CMV test results to UDOH (refer to **Option 3** at the bottom of the form). If the family refuses the CMV testing, please mark the appropriate box on the CMV and NBHS Testing Status FAX Form and fax this form along with the signed **CMV Testing Refusal Form** (also available on our CMV web site).

UDOH staff are here to help. To request a presentation on CMV hearing loss and/or UCA 26-10-10 for your facility or for questions or concerns, contact Stephanie McVicar at (801) 584-8218 or smcvicar@utah.gov.

The CPT code for CMV detection (qualitative) by PCR is 87496.

An appropriate ICD-9 code to put on the order is 389.8 (neonatal hearing loss).

The Growing Times

is a newsletter of the

Utah Chapter of the
American Academy of Pediatrics



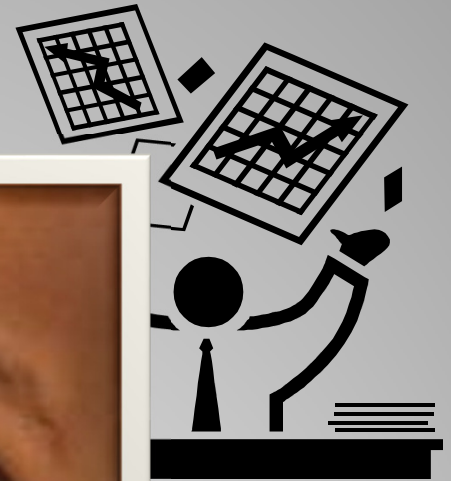
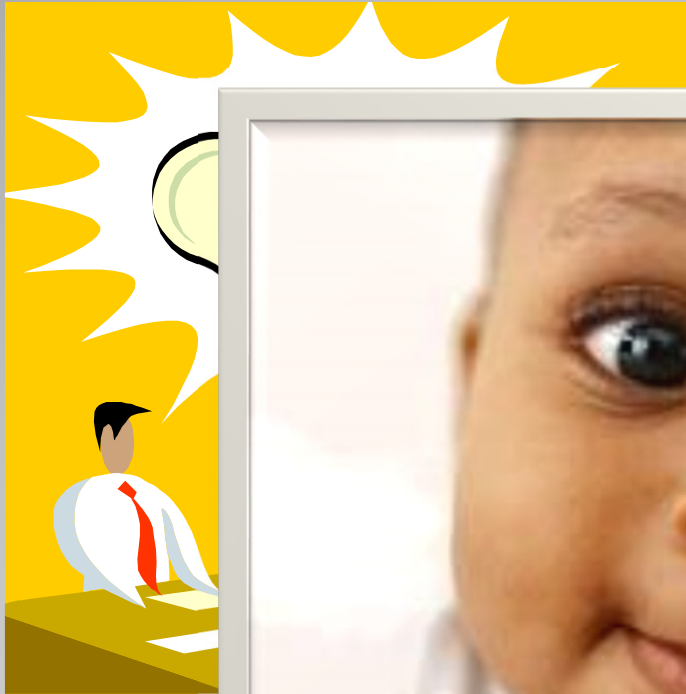
Share your comments!

We invite members to comment on current issues, articles, editorials, or submit story ideas. Deadline for submission of articles published in the next issue is March 20, 2014. Send to via email to office@aaputah.org.

Past editions of *The Growing Times* are available online at <http://www.aaputah.org>, click on Newsletter. For older editions, contact Cathy Oyler at office@aaputah.org or (801) 968-3411.



Need



Stakeholders





Photo: By Utah Images - Douglas Pulsipher

Ask Me
about CMV



Stephanie Browning McVicar, Au.D.,
CCC-A

smcvicar@utah.gov

(801) 584-8218

<http://www.health.utah.gov/cshcn/CHSS/CMV.html>



UTAH DEPARTMENT OF
HEALTH

www.health.utah.gov/cshcn/CHSS/CMV.html

Help

~~CMV~~

801-584-8215

QUESTIONS?